

ACUTE ILLNESS PROTOCOL
FATTY ACID OXIDATION DISORDERS
VERY LONG CHAIN ACYL CoA DEHYDROGENASE (VLCADD) DEFICIENCY

INTRODUCTION

Very long Chain Acyl CoA Dehydrogenase Deficiency (VLCADD) is an autosomal recessive disorder resulting in an intramitochondrial defect in the β -oxidation of fatty acids. It can cause severe hypoketotic hypoglycemia, encephalopathy, lethargy, liver dysfunction with hepatomegaly, cardiomyopathy metabolic acidosis, hyperammonemia and sudden death.